Journal of the American College of Cardiology
© 2010 by the American College of Cardiology Foundation
Published by Elsevier Inc.

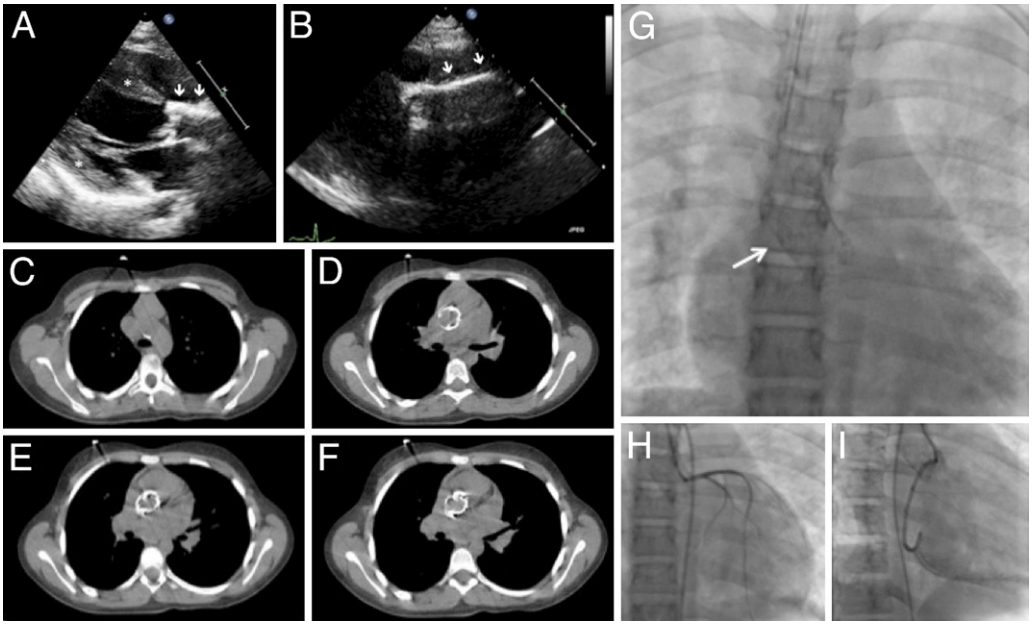
Vol. 56, No. 21, 2010
ISSN 0735-1097/$36.00
doi:10.1016/j.jacc.2010.02.078

IMAGES IN CARDIOLOGY

# Singleton-Merten Syndrome and Impaired Cardiac Function

Israel Valverde, MD,*† Eric Rosenthal, MD,* Aphrodite Tzifa, MD,*† Pravin Desai, MD,‡ Aaron Bell, MD,* Kuberan Pushparajah, MD,* Shakeel Qureshi, MD,* Philipp Beerbaum, MD,*† John Simpson, MD*

*London and Ipswich, United Kingdom*




From the *Department of Pediatric Cardiology, Evelina Children's Hospital, Guy's and St. Thomas Foundation Trust, London, United Kingdom; †Division of Imaging Sciences, King's College London, London, United Kingdom; and the ‡Child Health Department, Ipswich Hospital National Health Service Trust, Ipswich, United Kingdom.
Manuscript received February 19, 2010, accepted February 25, 2009.

A 13-year-old girl presented to our unit because of recurrent syncopal episodes. Echocardiography (**A and B**, Online Video 1) demonstrated left ventricular (LV) hypertrophy **(*)**, poor aortic valve excursion, and intense echogenicity extending to the ascending aorta **(arrows)**. Three-dimensional echocardiography showed dyskinesia of the LV with poor ejection fraction of 21%. The 32-slice computed tomography scan revealed the extension of the calcification up to the transverse arch **(C to F,** Online Video 2). The calcification was clearly delineated by fluoroscopy **(arrow)**, with normal coronary angiogram **(G to I)**.

The patient underwent aortic root replacement with pulmonary autograft. One week postoperatively, 3-dimensional echocardiography demonstrated dramatically improved LV ejection fraction (50%) with no evidence of dyskinesia (Online Video 3).

A diagnosis of Singleton-Merten syndrome, a rare genetic disorder characterized by calcium deposition, had been made recently. To our knowledge, cardiovascular imaging findings have not been reported in the literature.